Biopsy May Help Identify Early Pyoderma Faciale (Rosacea Fulminans)

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Pyoderma faciale is an uncommon acute presentation of rosacea. Edema, nodules, and draining sinuses may occur. Women in their early 20s are typically affected, and severe scarring may result in untreated cases. We report the case of a woman in whom a biopsy helped establish an early diagnosis so that decisive intervention could be initiated and scarring avoided. The histologic features of pyoderma faciale only rarely have been described and are illustrated in this case. Cutis. 2006;77:225-227.

yoderma faciale is an uncommon disorder that occurs mostly in postadolescent women. Rapid onset and absence of typical acne vulgaris lesions on the chest and back are helpful clues to the diagnosis. Contact dermatitis, angioedema, and acute lupus erythematosus may be considered in the clinical differential diagnosis because of the intense redness and swelling. Although its onset is usually idiopathic, pyoderma faciale is occasionally associated with Crohn disease.¹ Gram-negative bacteria such as Enterobacter cloacae and Klebsiella oxytoca sometimes can be cultured from lesions.² Histologic evaluation typically reveals a mixed periadnexal infiltrate with neutrophils, eosinophils, and granulomatous foci. The histiocytes in the infiltrate are reminiscent of the histologic findings that may be seen in early lesions of granulomatous cheilitis or Melkersson-Rosenthal syndrome. If the lesions are allowed to progress, deep abscesses, draining sinuses, and scarring result.3 Corticosteroids and isotretinoin have been useful therapeutic options.4-7 Performing an early biopsy in some cases can exclude other disease entities because of characteristic histologic findings.⁸

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Figure 1. Extensive facial erythema that is symmetrical and evenly distributed. Occasional papules, nodules, and small pustules are noted.

Case Report

A 37-year-old woman presented for evaluation of acute facial erythema and edema associated with tenderness and discomfort (Figure 1). The patient did not have fever or arthritis. Papules, nodules, and small pustules were noted. A 4-mm punch biopsy specimen taken from a left chin lesion revealed a superficial and deep perivascular and periappendiceal infiltrate consisting of lymphocytes, histiocytes, and occasional eosinophils (Figures 2 and 3). Neutrophils were noted within sebaceous lobules, and small collections of histiocytes were encountered (Figure 4). The histologic findings were consistent with pyoderma faciale (rosacea fulminans) and excluded contact dermatitis, angioedema, and acute lupus erythematosus. The

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Figure 2. Perivascular and perifollicular inflammation with no interface change or spongiosis (H&E, original magnification \times 10).

Figure 3. Inflammation centered around pilosebaceous units. The infiltrate consists of neutrophils, lymphocytes, and histiocytes (H&E, original magnification ×40).

Figure 4. Histiocytes and lymphocytes surround a follicle, even in early lesions (H&E, original magnification ×80).

	Perivascular Lymphocytic Infiltrate	Spongiosis	Basal Layer Vacuolopathy	Granulomatous Foci	Inflammation of Sebaceous Lobules
Acne fulminans	+	+/-	-	-	+
Acute lupus erythematosus	+	-	+	_	_
Angioedema	+	-	-	-	-
Contact dermatitis	+	+	_	_	-
Pyoderma faciale	+	_	_	+	+

Histologic Differential for Acute Facial Erythema*

patient had been placed on prednisone 60 mg/d by her referring physician; her therapy was continued as prednisone 20 mg 3 times daily for 7 days and then tapered. Minocycline hydrochloride 100 mg twice daily also was prescribed. She showed marked improvement within one week.

Comment

Early treatment of pyoderma faciale can help prevent scarring by preventing the formation of deep pustular lesions. The clinical differential diagnosis may include angioedema as well as other entities such as acute lupus erythematosus. Clinical diagnosis becomes easier when more established lesions are present, but the presence of more established lesions correlates with increased scarring and slower recovery. A biopsy result showing a perivascular and perifollicular infiltrate of lymphocytes and histiocytes without interface change or spongiosis allows for diagnosis and helps to exclude the other disorders in the differential diagnosis with confidence (Table). The prominent involvement of sebaceous lobules is characteristic. Histiocytes present in early lesions also are typical. Early treatment with systemic corticosteroids has a marked impact, with rapid improvement of pain, swelling, and erythema. Systemic antibiotics often are included in the treatment plan for acne fulminans and pyoderma faciale.⁹ Follow-up treatment with oral isotretinoin at a dose of 1 mg/kg daily for a 16- to 20-week course allows for long-term control of the disease.

The presence of histiocytes in the inflammatory infiltrate and the histologic resemblance to early granulomatous cheilitis is interesting because both entities have been associated with Crohn disease.^{3,9} Other severe and extensive acneform lesions, such as

those seen with acne fulminans, can be associated with joint pain, fever, and even erythema nodosum.¹⁰ The intense neutrophilic infiltrate in these disorders and some of their common associations are reminiscent of pathergy and suggest some similarities in their underlying inflammatory pathways.

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